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CASE REPORT

Metastatic Synovial Sarcoma With Cervical Spinal Cord Compression Treated With Posterior Ventral Resection: Case Report

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Abstract

Context: Synovial sarcomas, which represent 5% to 10% of all adult soft-tissue sarcomas, usually metastasize to the lungs. Metastasis to the spine is rare. Spinal cord compression due to spinal metastasis occurs in approximately 3% of patients with extraspinal soft-tissue sarcomas.

Method: Case report.

Findings: A 26-year-old woman presented with neck pain, arm weakness, and a history of metastatic synovial sarcoma originating at the right knee. Computed tomography revealed destruction of the odontoid and C2 body. Magnetic resonance imaging revealed tumor in the posterior elements of C2 and in the ventral epidural space from C2–C5. She was treated with C2–C3 laminectomy, posterior C2 corpectomy with occipital-C7 fixation, and fusion. Postoperatively, her neck pain resolved and left upper extremity strength returned to normal.

Conclusion and Clinical Relevance: Metastatic spinal cord compression from synovial sarcoma is rare. Surgical resection can lead to neurologic improvement.

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Key Words: Sarcoma, synovial, metastatic; Cervical vertebrae, neoplasia; Spinal fusion; Spinal cord, compression; Tumor, spinal, epidural

INTRODUCTION

Synovial sarcomas are rare tumors, representing 5% to 10% of all adult soft-tissue sarcomas (1). Predisposing etiologic agents or genetic conditions have not been identified. Synovial sarcomas can arise from almost any anatomic site, but the extremities are the most common sites of primary involvement. The most common non-extremity sites of primary disease include the trunk, retroperitoneal/abdominal region, and head and neck (1,2). The vast majority of primary synovial sarcomas metastasize to the lungs (1,2). Metastasis to the spine is rare. Spinal cord compression due to spinal metastasis occurs in approximately 3% of patients with extraspinal soft-tissue sarcomas, with the lumbar-sacral spine being the most common level, followed by the thoracic spine and then the cervical spine (3). We report a rare case of metastasis to the cervical spine.

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CASE REPORT

A 26-year-old woman presented with neck pain and left arm weakness. Past history was significant for metastatic synovial sarcoma. At approximately age 10 years, while living in Mexico, the patient underwent resection of synovial sarcoma of the right knee. Three additional resections were performed for 3 subsequent recurrences. At approximately 18 years of age, the sarcoma recurred for the fourth time and was resected. The treatment was followed by radiation. Shortly after, the patient immigrated to the USA. Five years later, the tumor again recurred in the right knee and right thigh. The patient underwent 7 cycles of chemotherapy and for the next 20 months did well without any symptoms. Then a mass was found in the lung and resected. Pathologic evaluation was consistent with monophasic synovial sarcoma.

Ten weeks after resection of the lung mass, the patient was informed that she had widely extensive metastatic disease. Four weeks later, she presented to our service with neck pain and left arm weakness. Neurologic examination revealed motor strength to be 5/5 in all muscle groups of the upper and lower extremities, with the exception of the left upper extremity. She was found to have 3/5 left deltoid strength, 3/5 left biceps, 4+/5 left

triceps, and 5/5 left hand grip strength. The rest of the neurologic examination was normal.

Computed tomography of the cervical spine showed lytic destruction of the dens and body of C2 with narrowing of the spinal canal and compression of the cord at that level. There was also an adjacent soft-tissue component extending into the prevertebral soft tissues, and a pathologic fracture with complete disassociation of the dens from the body of C2 without significant displacement (Figure 1A and B). Computed tomography of the thoracic spine revealed lytic lesions involving the T4 vertebral body centrally, as well as the right aspect of the T6 and T7 vertebral bodies and the posterior T8 and T11 vertebral bodies.

Magnetic resonance imaging of the cervical spine revealed a mass replacing most of the C2 vertebral body (Figure 2A and B). The enhancing soft tissue extended into the prevertebral space anterior to C2 and C3. There was an additional bilateral paravertebral component extending throughout C2 and C3. There was also extension on the posterior aspects of the C2–C4 vertebral bodies with an epidural component identified. The epidural tumor caused marked narrowing of the spinal canal to approximately 5 mm and also exerted a mass effect on the cord. The left vertebral artery was not well visualized at the level of the mass between the C2 and C4 levels, which was suspicious for vertebral artery occlusion.

Preoperative staging of the neoplasm was not necessary because widely metastatic disease had been confirmed just 1 month before consultation. Because of the extent of the ventral tumor, the patient underwent a C2–C3 laminectomy, posterior C2 corpectomy with occipital-C7 fixation, and allograft fusion. After the laminectomy was performed, the pars and pedicles were resected bilaterally, allowing access to the ventral epidural space. The tumor was then removed piecemeal and the spinal cord was decompressed. Postsurgical radiographs of the cervical spine (Figure 3) revealed the resection of the C2 vertebral body. Postoperatively, the patient's left upper extremity strength returned to normal. Due to the poor prognosis and known metastatic disease, complete resection by an anterior approach was not attempted.

Pathologic evaluation of both the soft tissue and bone revealed malignant spindle cell tumor consistent with metastatic sarcoma, primarily synovial with some areas associated with a slight myxoid to lipoblastic appearance. The histologic appearance of the tumor was essentially the same as that seen in previous sections from the knee and lung; the tumor was composed of mitotically active spindle cells arranged in a fascicular pattern, with no epithelial component identified, typical of monophasic synovial sarcoma (Figure 4A and B).

The patient did well postoperatively and was transferred to the medicine service several days postoperatively, where she continued to improve (Figure 5). Physical therapy was continued as tolerated. One month after surgery, the patient was to undergo palliative

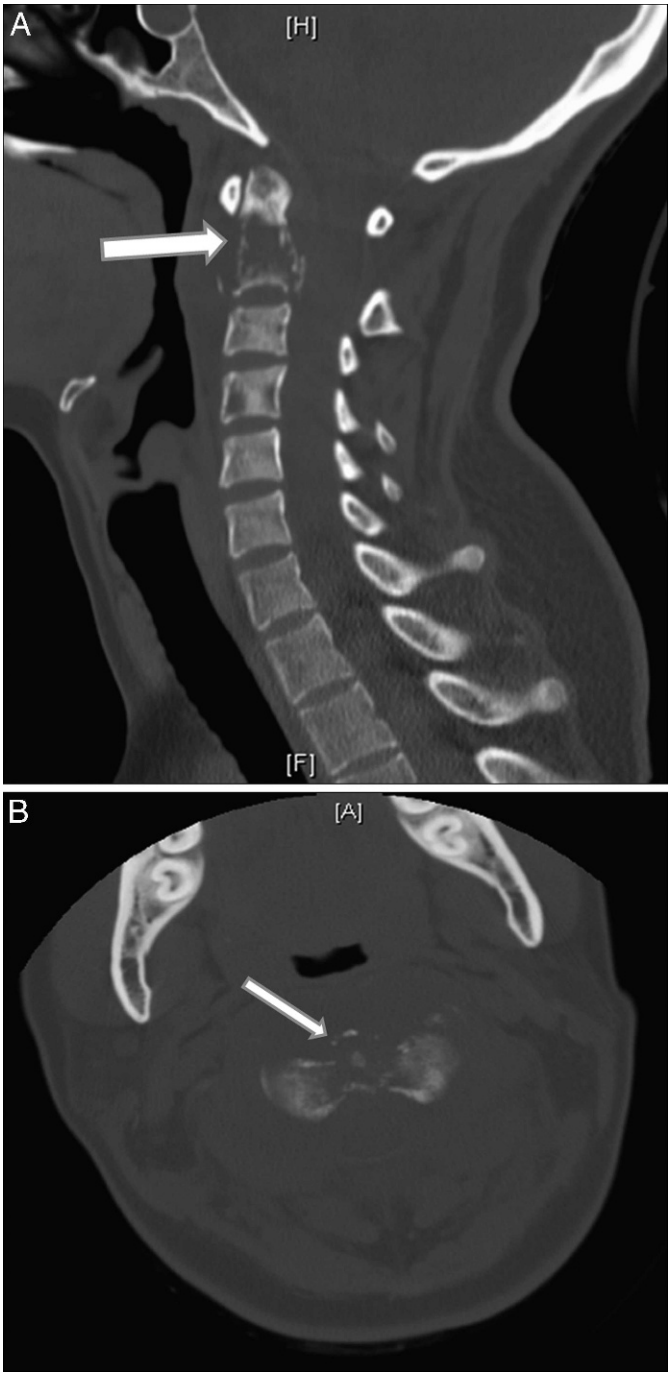
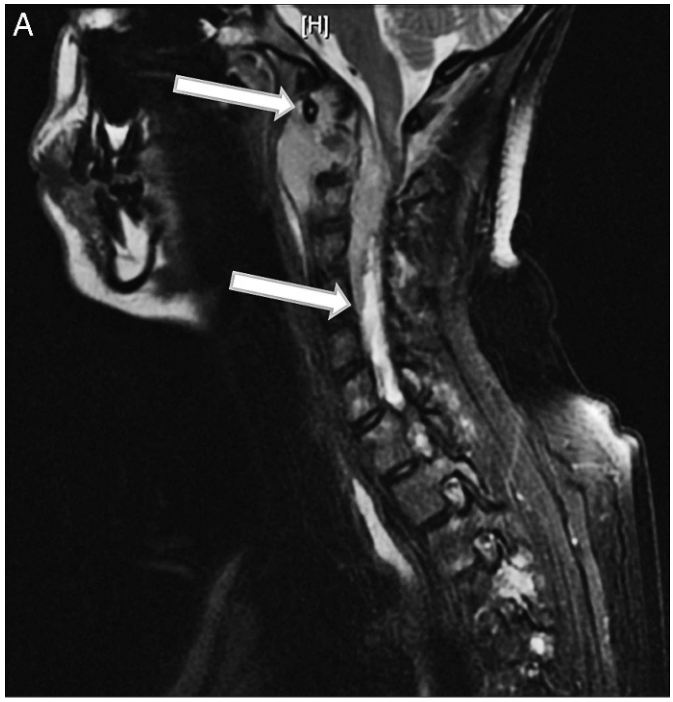


Figure 1. *(A) Sagittal reconstruction computed tomographic scan of the cervical spine showed permeative lytic destruction of the dens and body of C2 with narrowing of the spinal canal and compression of the cord at that level. Also noted was an adjacent soft-tissue component extending into the prevertebral soft tissues and a pathologic fracture with complete disassociation of the dens from the body of C2 without significant displacement (arrow). (B) Axial computed tomographic scan of the cervical spine showed destruction of the odontoid with a pathologic fracture through the base of the dens.*



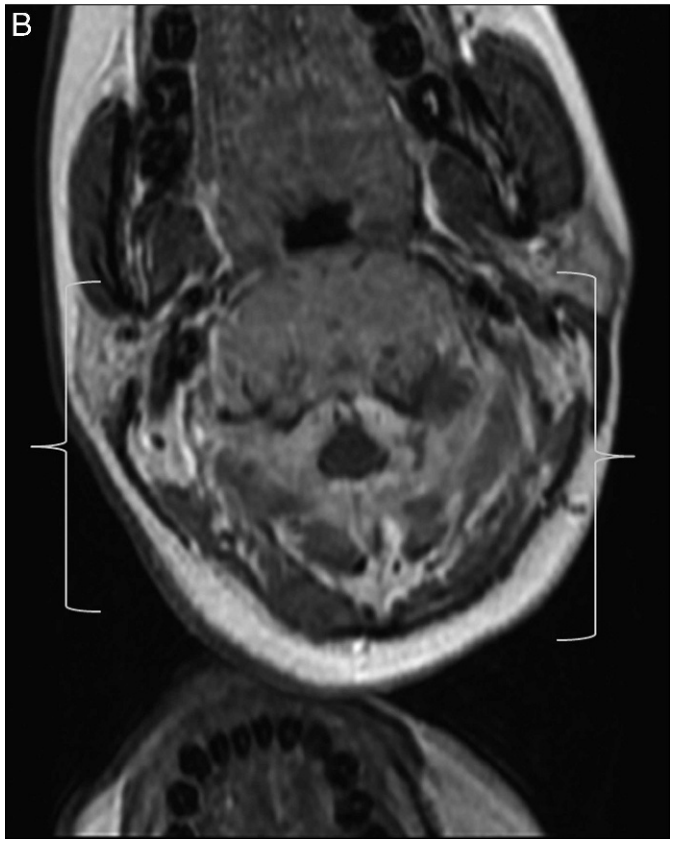


Figure 2. *(A) Sagittal T2W1 magnetic resonance imaging revealed a mass replacing most of the C2 vertebral body. There was involvement of the odontoid and ventral epidural space from C2–C5. (B) Axial T1W1 magnetic resonance image shows circumferential tumor at C2 compressing the spinal cord.*

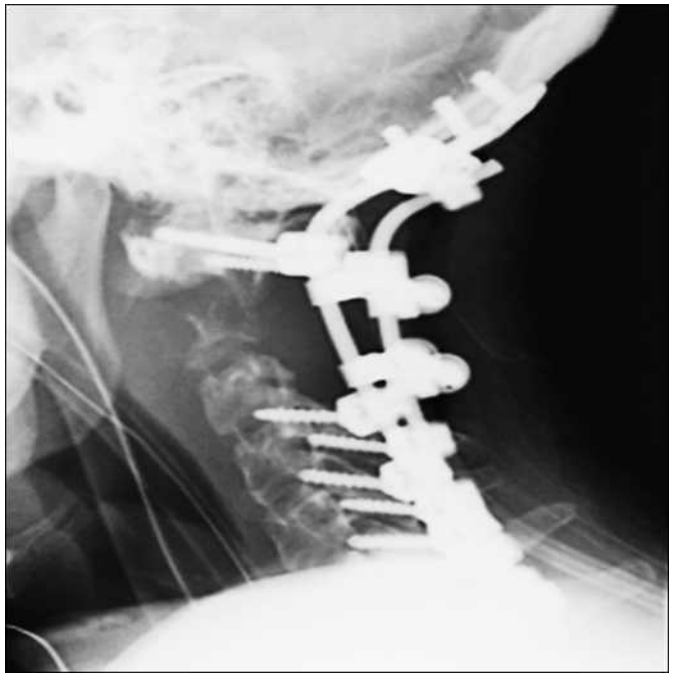


Figure 3. *Postsurgical lateral radiograph of the cervical spine revealed the resection of the C2 vertebral body and confirmed the placement of posterior spinal fixation with posterior rods extending from the occipital bone to the lower cervical spine with lateral mass screws identified at multiple levels.*

chemoradiation but developed fever, leukocytosis, and acidosis and ultimately became septic. Computed tomographic scans showed multiple metastases to the liver and abdominal parenchyma. She was no longer eligible for systemic chemotherapy and radiation therapy due to the heavy tumor burden. She died 6 months postoperatively of disease progression.

DISCUSSION

Synovial sarcomas are rare tumors with an incidence of approximately 2.75 cases per 100,000 people (4) and represent only 5% to 10% of all adult soft-tissue sarcomas and 12% to 15% of all adult extremity soft-tissue sarcomas (1). Synovial sarcoma is the third most common adult extremity soft-tissue sarcoma (1). Synovial sarcoma accounts for approximately 1% of all childhood malignancies and, despite its rarity, is the most common nonrhabdomyosarcoma in childhood, accounting for approximately 30% of all soft-tissue sarcomas (5–7). Synovial sarcomas are considered high-grade soft-tissue sarcomas, in contrast to most other soft-tissue sarcomas that tend to have both high- and low-grade versions (1). Growth rate is slow in its early stages but increases rapidly over the years.

Synovial sarcomas can arise from any anatomic site, but the para-articular regions of the extremities are the most common site of primary involvement. They almost never arise within the joint, and they are not associated

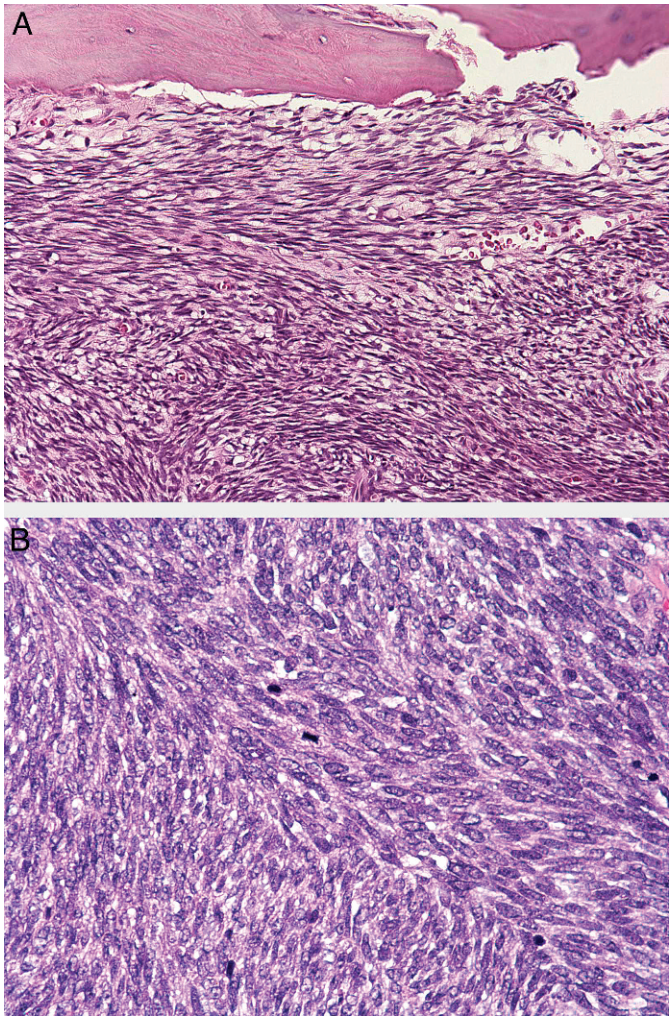


Figure 4. *(A and B). Biopsy of the recurrent right knee mass at age 23 years reveals a tumor composed of mitotically active spindle cells arranged in a fascicular pattern, with no epithelial component identified, typical of monophasic synovial sarcoma. The cells were immunoreactive with vimentin and CD99 (hematoxylin-eosin, original magnification ×400).*

with normal synovial tissue; the cell of origin has been determined to be epithelial (1). Only about 20% of all primary synovial sarcomas arise in nonextremity sites, the most common of which are the trunk (~8%), retroperitoneal/abdominal sites (~7%), and head and neck (~5%) (1,2).

The presentation of synovial sarcomas is generally classified as monophasic or biphasic. Monophasic synovial sarcomas are composed entirely of ovoid/spindle cell morphology, whereas biphasic subtypes are composed of both spindle cell and epithelial cell components. Recently, a third and rare poorly differentiated subtype of synovial sarcoma has been described, which is composed of uniform, densely packed, small ovoid blue cells that resemble other small round blue cell tumors (1). Poorly differentiated synovial sarcomas present a diagnostic

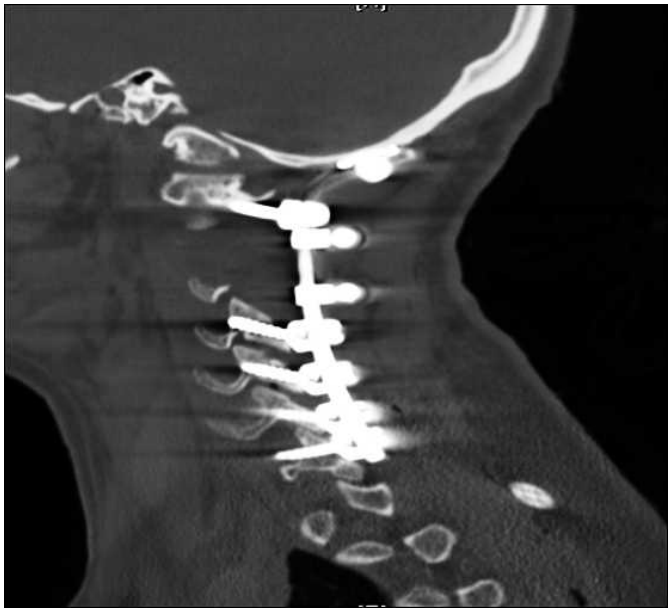


Figure 5. *Two weeks postoperatively, computed tomography of the cervical and thoracic spine revealed occipital and posterior spinal fusion with screws in the skull base, C1, and C4–C7. The C2 body has been resected.*

challenge because they can mimic many tumors, including poorly differentiated carcinoma, Ewing's/primitive neuroectodermal tumor, and epithelioid fibrosarcoma (1). Because synovial sarcoma can be very difficult to diagnose by histology and immunohistochemical results alone, highly sensitive and specific tests have been developed for use on routinely processed, formalin-fixed, paraffin-embedded tissues. If diagnostic confirmation still cannot be obtained, molecular testing should be performed (1).

Prognostic factors for disease-specific survival in synovial sarcoma include age, primary tumor size, margin of resection, mitotic activity, bone or neurovascular invasion, histologic subtype, overexpression of the cellular tumor antigen p53, the rate of cell proliferation (as assessed by Ki-67 immunoreactivity, the Ki-67 index of cellular proliferative activity), and the gene fusion transcript (SYT-SSX) fusion type (1,2,8,9). Of all these factors described, the relative predictive value remains controversial; however, large tumor size has consistently proved to be associated with development of distant metastasis and decreased disease-specific survival (1,8,10).

The recurrence rate of synovial sarcoma is 28% to 70% (4). Tumors usually recur within 2 years of initial therapy, but there are reports of recurrence after 30 years (4). In a study by Singer et al (9), almost 50% of the deaths observed occurred from 5 to 10 years after initial diagnosis, emphasizing the relatively high incidence of late metastasis and the need for long-term follow up. The vast majority of primary synovial sarcomas metastasize to the lungs (1,2). Metastases occur in approximately half of all cases of synovial sarcomas (1,4,8), with a resultant 5-year disease-specific mortality of 37% (8). The sites most

affected by metastases are the lung, lymph nodes, and bone marrow (4).

Metastasis of synovial sarcoma to the spine is extremely rare, and data in the literature consist primarily of case reports. Spinal cord compression due to spinal metastasis, an emergent oncologic event, occurs in approximately 3% of patients with extraspinal soft-tissue sarcomas, with the lumbar-sacral spine being the most common level (42%), followed by the thoracic spine (46%) and the cervical spine (12%) (3). After the diagnosis of spinal cord compression in soft-tissue sarcomas, the survival rate is 1 to 21 months, with a median rate of 5 months (3). There is only 1 reported case of synovial sarcoma originating from the lumbar spine (4) and few cases of metastasis to the spine (3,11–14). In a series of 59 patients, Bilsky et al (11) noted 3 spindle-cell sarcoma metastases to the spine but did not specify the levels. In a series of 19 patients, Merimsky et al (3) reported 3 patients with spinal metastases, to T7–T8, C2–C3, and S1–S3. Three separate case reports documented metastasis to the lumbar vertebra (12,13) and to the thoracic vertebra (14).

Metastatic synovial sarcoma conveys a poor prognosis. In a study of 271 patients with synovial sarcoma, 16 developed metastases. Metastases affected the lung in 14 cases, bone and lung in 1 case, and the liver in 1 case. Fourteen patients died of disease at a median of 18 months from diagnosis, and 2 were alive with progressive disease at the time of the report (2).

If possible, complete surgical resection of primary synovial sarcoma remains the treatment of choice (1,15). Adjuvant radiation therapy after surgery provides excellent local control (15,16). Response to chemotherapy is highly objective. However, one study showed no detectable beneficial effects on survival in the subset of patients treated with chemotherapy vs those who received no chemotherapy (9). Other studies showed an improvement in metastasis-free survival in patients treated with chemotherapy (2,8). Local recurrences can be treated with limb-sparing surgery. Due to this patient's widely metastatic disease, complete resection was not performed in this case.

CONCLUSION

Although metastasis to the spine from synovial sarcoma is extremely rare, it should be considered in the differential diagnosis in patients who have synovial sarcoma and a spine lesion. Patients who present with this tumor and concomitant back pain and neurologic symptoms should have a thorough radiologic workup, including computed tomography and magnetic resonance imaging, to confirm the diagnosis and ensure appropriate management. Surgical resection can lead to neurologic improvement in patients who present with spinal cord compression.

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